Making Sense of Complex Repairs 20 years Later

Mark Twite
MA MB BChir FRCP
Associate Professor,
Department of Anesthesiology
Director of Pediatric Cardiac Anesthesia
Children’s Hospital Colorado
University of Colorado
Anschutz Medical Campus

Objectives

1. Identify the anatomy of complex congenital heart repairs
2. Differentiate uncomplicated from complicated repairs with respect to physiology and anesthetic management
3. Discuss Single ventricle, TGA and Eisenmenger syndrome physiology and anesthetic management

ACHD

• CHD 5 - 9 of 1,000 live births
  – 1.5 of 1,000 live births have complex CHD
• More adults than children live with CHD in USA
  – ~1 - 3 million adults with CHD in USA & Canada
  – ~1.8 million adults with CHD in Europe
• Improvement in survival over the past 20 years
  – 90% of children survive to adulthood
• USA estimates 500,000 adults with complex CHD
  – Only 10% receive follow-up care in a ACHD center

What is so unique about ACHD population?

• Society has already invested a large amount of resources to achieve survival to adulthood
• Young adults with CHD have the potential to contribute to the GDP for 30-40 years
• The period of early adulthood is relatively uneventful in terms of complications and resource utilization compared with early childhood and later adult life

However……..

• Many young adults with ACHD do not receive cardiology follow-up
  – Re-location with school and work
  – Health insurance
  – Perception that they are doing well
  – Lack of transition from pediatric to adult programs
• This lack of preventive care may increase the overall costs of care
Perioperative Outcomes

- Major non-cardiac surgery in ACHD
  - Greater morbidity and mortality
  - ACHD independent predictor of increased mortality
- Vulnerable population
  - 50% of adults with CHD can not correctly name or describe their diagnosis
  - Majority of anesthesia providers do not have the knowledge and are not comfortable looking after patients with ACHD, especially as complexity increases

Anesthesia providers and ACHD

- ACHD patients presenting for non-cardiac surgery
- Highest knowledge and comfort scores for:
  - Fellowships in cardiac anesthesia and pediatric anesthesia
  - Increased frequency of CPB cases
  - Increased frequency of providing care for patients under 2yrs of age
- Implications for training

Increasing non-cardiac surgery in ACHD patients

![Graph showing increasing non-cardiac surgery in ACHD patients over time](Maxwell 2013)

Mortality by lesion type

<table>
<thead>
<tr>
<th>Lesion Type</th>
<th>n (%)</th>
<th>Died</th>
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<tbody>
<tr>
<td>Atrial septal defect</td>
<td>4,908</td>
<td>156 (3.2)</td>
</tr>
<tr>
<td>Congenital aortic</td>
<td>1,789</td>
<td>53 (3.0)</td>
</tr>
<tr>
<td>stenosis/aortic insufficiency</td>
<td>85</td>
<td>3 (3.5)</td>
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<tr>
<td>Congenital mitral</td>
<td>409</td>
<td>10 (2.1)</td>
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<tr>
<td>stenosis/regurgitation</td>
<td>248</td>
<td>9 (3.6)</td>
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<tr>
<td>Congenital coronary anomaly</td>
<td>339</td>
<td>13 (3.8)</td>
</tr>
<tr>
<td>Tetralogy of Falot</td>
<td>121</td>
<td>7 (5.8)</td>
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<tr>
<td>Ventricular septal defect</td>
<td>831</td>
<td>52 (6.3)</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>65</td>
<td>4 (6.2)</td>
</tr>
<tr>
<td>Others</td>
<td>1,745</td>
<td>76 (4.4)</td>
</tr>
<tr>
<td>Combined complex†</td>
<td>544</td>
<td>35 (6.5)</td>
</tr>
</tbody>
</table>

Single Institution Data

Mayo Clinic, 2013

- All patients who had undergone Fontan palliation (n = 1,133)
- Patients > 16yrs, Fontan, for non-cardiac surgery
- 39 GAs given to 31 patients
  - 31% had perioperative complications
  - One death

![Mortality by lesion type table](Maxwell 2013)
ACHD: Anesthetic Considerations
- Detailed knowledge of anatomy & physiology
- Multidisciplinary team
- Increased perioperative risk
  - CHF
  - Pulmonary Hypertension
  - Cyanosis
  - Bleeding & thrombosis
  - Dysrhythmias

ACHD and CHF
CHF defined as:
- \( VO_2 < 25 \text{ml/kg/min} \) & NT-pro-BNP > 100pg/ml
  - 26% of ACHD, mostly young 30-40yrs
  - Increases as lesion complexity increases
- Greatest risk of CHF:
  - Single ventricle (R > L)
  - Tetralogy of Fallot s/p repair with PI
  - TGA

Probability of Heart Failure by CHD

Relative Risk for Specific Arrhythmias in Common Congenital Heart Defects

Complex Lesions and Physiology
- Single ventricle palliated to Fontan physiology
- Transposition of the Great Arteries (TGA)
- Eisenmenger syndrome
Single Ventricle Physiology
“So in whatsoever creature there is lungs, there is likewise in them two ventricles of the heart, the right and the left”
William Harvey 1638

However…..

1940s recognition that across species, PAP was 25/10 mmHg and venous pressure alone may be sufficient to move blood through the lungs

ACHD: Single Ventricle Physiology
• Palliative path for a univentricular heart
  – Stage 1: BT shunt, Norwood procedure, Sano shunt, PA Banding
  – Stage 2: Glenn shunt
  – Stage 3: Fontan
• The single ventricle may be morphologic left or right

Stage 1 Too much PBF
Tighten to PA Band to ½ systemic BP (Trusler’s rule) and SaO2 of 80%

Stage 1 Too little PBF
Classic BT shunt  Modified BT shunt
Establish a reliable source of PBF - balance Qp:Qs with Sats 80%

Stage 1 Norwood
BT-Shunt  Sano Modification
**Qp/Qs**

\[ \frac{Qp}{Qs} = \frac{(SaO_2 - SvO_2)}{(SpvO_2 - SpaO_2)} \]

- Assume SpvO_2 = 100%
- Measure SaO_2 = SpaO_2 = 80%
- Measure SvO_2 = 60%

\[ \frac{Qp}{Qs} = \frac{(80 - 60)}{(100 - 80)} = \frac{20}{20} = 1/1 \]

**Stage 2 Glenn Shunt**

- 2-6 months of age (PVR has decreased)
- Decrease volume load on the heart
- Sats 80%
- Physiology of PBF
- Pulmonary arterial venous AVMs develop due to lack of hepatic factor to lungs
- Monitoring issues

**Stage 3: Fontan**

- 2-6 months of age (PVR has decreased)
- Decrease volume load on the heart
- Sats 80%
- Physiology of PBF
- Pulmonary arterial venous AVMs develop due to lack of hepatic factor to lungs
- Monitoring issues

**Fontan Fluid Dynamics**

- Preserve fluid energy
- Preserve normal atrial pressures
- Eliminates extensive atrial suture lines (decrease arrhythmias)
- Fenestration allows systemic preload to be maintained (at the expense of saturation) in the event of increased PVR. May be closed in cath lab at a later time.

**Fontan Physiology and PBF**

1. Spontaneous Ventilation
   - Increased venous return and PBF
   - BUT avoid hypercarbia, hypoxia, atelectasis and acidosis
     - Increase PVR
     - Decrease PBF and CO
Fontan Physiology and PBF

2. Positive Pressure Ventilation
   • PBF during the expiratory phase
     – Limit PIP < 20 cmH₂O
     – Low RR (<20 bpm)
     – Short inspiratory times
     – Avoid high PEEP (but avoid atelectasis)
     – Tidal volumes 10ml/kg
     – Early extubation
   • Adequate intravascular volume

Optimizing Ventilation

Prevent an increase in PVR

Fontan Failure

1. Cardiac
   • Arrhythmias
   • Congestive heart failure (Increased work of single ventricle)
     – Problems with ECHO estimations
   • AV valve regurgitation

Fontan Arrhythmias

Fontan Failure

2. Pulmonary
   • Increasing PVR
   • Cyanosis
     – Pulmonary AVMs, fenestration
   • Pleural effusions
   • Plastic bronchitis
Plastic Bronchitis

Fontan Failure

3. Hepatic
   • Dysfunction
     – Synthetic function decreased
   • Protein-losing enteropathy
     – Loss of proteins, immunoglobulins
     – Ascites
   • Esophageal varices

4. Hematologic
   • Thromboembolic
     – Hypercoagulability, atrial arrhythmias
     – Passive venous flow
   • Often on aspirin and/or Coumadin

Fontan: Perioperative decreased CO

• Hypovolemia (NPO status)
• Positive pressure ventilation
• Hypercarbia
• Hypoxemia
• Increase venous capacitance (anesthetics)
• Ventricular dysfunction
• Arrhythmias
• Increase PVR (effusion, ascites, hypothermia, pain)

Fontan: Periop strategies to increase CO

• Optimize ventilation
  – FiO₂ 1.0, pCO₂ 30 mmHg, pH 7.45
  – Consider iNO 20-40ppm
• Adequate anesthesia & analgesia
• Normothermia
• Inotrope support
  – Milrinone
  – Dobutamine

Future Fontan
Complex Lesions and Physiology

- Single ventricle palliated to Fontan physiology
- Transposition of the Great Arteries (TGA)
- Eisenmenger syndrome

Anatomy of TGA

- Atrio-ventricular concordance; ventriculo-arterial discordance
- d-TGA
  - Aorta is anterior and to the right of the pulmonary trunk (as opposed to posterior)
  - Refers to embryological looping

TGA: diagnosis in fetal life

- Chronic cerebral hypoxia – delayed brain maturation
- Antenatal diagnosis (75% in modern centers) has a beneficial effect on pre-op status and post-op outcome
- High pre-op lactate predicts poor neurological outcome

Types of TGA

1. TGA with intact ventricular septum (TGA/IVS)
   - 85% of cases
2. TGA with ventricular septal defect (TGA/VSD)
   - 10% of cases
   - Associated with other abnormalities: right side aortic arch, IAA, coarctation of aorta
3. TGA/VSD with left ventricular outflow tract obstruction (TGA/VSD/LVOTO)
   - Abnormal pulmonary valve not suitable for arterial switch operation (ASO)
4. Congenitally corrected TGA (ccTGA)

Coronary artery anatomy

- The coronary arteries will need to be moved during the ASO
- Anomalies of the coronary arterial course which are most challenging:
  - Intramural
  - Coronary artery stretches over RVOT
  - Lying close to a commissure

TGA: early management

- Balloon atrial septostomy (BAS) (Miller & Rashkind 1964) may predispose to cerebral embolism but is needed in the majority of infants
- Failure of cyanosis to resolve after PGE and BAS may indicate pulmonary hypertension (12%)
Taussig-Bing malformation

1949 Helen Taussig and Richard Bing
• DORV with both great vessels from the RV
• VSD streams blood from LV to PA and RV to Aorta
• Repair by tunnelling VSD to PA (create TGA) and then perform ASO (undo the TGA)

TGA: options for surgical correction

• 1959 Senning procedure
  – Complex re-routing of blood in the atria
• 1964 Mustard procedure
  – Simpler atrial baffling
  • Right blood into the right great artery but from the wrong ventricle
  • Problems with:
    – Conduction system
    – Baffle leaks and obstruction
    – RV failure
    – Systemic (tricuspid) AV valve failure

TGA: Senning-Mustard Repair

• 1975 Adib Jatene performed the first ASO in Brazil
• True anatomical correction of TGA

Arterial Switch Operation (ASO)

Lecompte Maneuver: the PAs are brought anterior to the aorta

TGA/VSD/LVOTO

• 1969 Giancario Rastelli Mayo Clinic
• Pulmonary valve is no good so precludes the ASO
• Rather than closing the VSD it is baffled to the aorta and an RV-PA homograft is placed (requires replacement)
Rastelli Procedure

ccTGA

- Double discordance
  - Atrioventricular
  - Ventriculoarterial
- Physiologically corrected transposition
- L-TGA
- High incidence of Ebstein like dysplasia of the systemic (tricuspid) AV valve

ccTGA

- Often survive to age 50yrs uncorrected BUT
  - Develop AV valve regurgitation
  - Impaired RV function
  - Rhythm disturbance
- Classic repair
  - Fix any intracardiac defects such as VSD, repair tricuspid (systemic) AV valve
  - Pacemaker placement
  - Leave the RV systemic

The double switch for ccTGA

1. ASO
2. Senning-Mustard

The double switch for ccTGA

- What if there is LVOTO?
  1. Senning – Mustard
  2. Rastelli

Outcomes of ASO

- Survival approaching 100%
  - Mild developmental delay
- Pulmonary artery stenosis
  - Re-operation or cardiac cath in 30% of long term survivors
  - Be careful with pulmonary artery stents: compression of aorta and/or coronary artery
Late complications: adults after ASO

1. Arrhythmias
   - More common in baffle procedure
   - 10% including AVN block, SVT and VT (increased with VSD)

2. Coronary artery dysfunction
   - Clinically silent ischemia due to denervated heart
   - Usually present with pallor, VT or sudden death
   - Significant problems in 10% patients so far
   - Screening

3. Dilation of the neo-aortic root and AI

Complex Lesions and Physiology

- Single ventricle palliated to Fontan physiology
- Transposition of the Great Arteries (TGA)
- Eisenmenger syndrome

Eisenmenger Syndrome

- Victor Eisenmenger 1897
  - 32yr old man with cyanosis and hemoptysis
  - Large VSD on post-mortem

- Paul Wood 1958
  - Coined the term Eisenmenger syndrome
  - Presence of a congenital heart defect permitting increased pulmonary blood flow/pressure resulting in increased PVR and reversed shunt

CHD and the likelihood of developing PH if not repaired within the designated time frame

- Truncus arteriosus 100%
- AVC 100%
- TGV 100%
- Large VSD: 50%
- Large PDA: 50%
- Large ASD: 10%

Pre-tricuspid shunt (increase in flow) and post-tricuspid shunt (increase in flow and pressure)

Development of Eisenmenger Syndrome

- Left-to-right shunt
- Increased pulmonary blood flow (shear stress/endothelial stretch)
- Endothelial dysfunction and vascular remodeling
- Smooth muscle cell proliferation, increase in extracellular matrix, intravascular thrombosis
- Increase in PVR
- Inverted shunt: right-to-left
- Cyanosis (Eisenmenger syndrome)

Clinical manifestations of ES

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>Clinical manifestation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eisenmenger</td>
<td>Fatigue, dyspnea, syncope, sudden death</td>
</tr>
<tr>
<td>Secondary erythrocytosis</td>
<td>Polycythemia, right ventricular hypertrophy, cyanosis</td>
</tr>
<tr>
<td>Renal insufficiency</td>
<td>Proteinuria, elevated creatinine, hypertension</td>
</tr>
<tr>
<td>Acute right ventricular failure</td>
<td>Heart failure, shock</td>
</tr>
<tr>
<td>Interstitial lung disease</td>
<td>Interstitial lung disease, hypoxemia, hypoxemia, respiratory failure</td>
</tr>
<tr>
<td>Right ventricular dilatation and right heart failure</td>
<td>Right heart failure, cyanosis</td>
</tr>
<tr>
<td>Renal dysfunction</td>
<td>Increased blood urea nitrogen, hypoxemia, respiratory failure</td>
</tr>
<tr>
<td>Hepatic venous congestion</td>
<td>Hepatic congestion, ascites, peripheral edema</td>
</tr>
<tr>
<td>Infections</td>
<td>Endocarditis, cerebral abscess</td>
</tr>
<tr>
<td>Isoimmune hemolytic anemia</td>
<td>Hemolytic anemia, hyperbilirubinemia, jaundice, anemia</td>
</tr>
</tbody>
</table>

Rosenzweig 2012

Kumar 2009
Treatment of ES

1. Pharmacological
   A. Digoxin, diuretics, anticoagulants
   B. PAH pathways
      i. Treat and repair?
2. Phlebotomy
3. Transplantation
   A. Lung Tx with correction of cardiac defect
   B. Heart-Lung Tx

BREATHE-5
Bosentan Randomized Trial of ERA Therapy-5

Eisenmenger: Treat and Repair?

Conclusions

• Increasing number of adult survivors with complex CHD
• Perioperative care may be in smaller centers with little pre-op information
• Understand the plumbing!
• Consider CHF, arrhythmias, pulmonary hypertension, bleeding/thrombosis, and cardiopulmonary interactions

Thankyou!
References